

## Case Report

# Intraventricular Hemorrhage Long after Successful Encephaloduroarterio Synangiosis in Moyamoya Patient

Moon Young Chung, M.D.,<sup>2</sup> Young Seok Park, M.D.,<sup>1</sup> Dong-Seok Kim, M.D., PhD.,<sup>1</sup> Joong-Uhn Choi, M.D., PhD.<sup>2</sup>

Department of Neurosurgery,<sup>1</sup> Severance Children's Hospital, Brain Korea 21 Project for Medical Science, Brain Research Institute, Yonsei University College of Medicine, Seoul, Korea

Department of Neurosurgery,<sup>2</sup> Institute for Clinical Research, College of Medicine, CHA Medical University, Seongnam, Korea

Intraventricular hemorrhage long after successful encephaloduroarterio synangiosis (EDAS) is very rare. The effect of revascularization surgery for preventing hemorrhagic event of moyamoya disease remains controversial. We report a 17-year-old female with intracerebral hemorrhage and intraventricular hemorrhage 10 years after successful EDAS. Even though cerebral vessels angiography showed good collateral circulations without specific weak points, a cerebral hemorrhage could occur in patient with ischemic type of moyamoya disease long after successful indirect bypass operations. Good collateralization of cerebral angiography or magnetic resonance perfusion image after indirect bypass surgery would ensure against ischemic symptoms, not a hemorrhage. And, thus a life-time follow-up strategy might be necessary even if a good collateral circulation has been established.

**KEY WORDS :** Moyamoya disease · Intracranial hemorrhage · Perfusion · Ischemic attack

## INTRODUCTION

Moyamoya disease is an unusual form of chronic, occlusive cerebrovascular disease characterized by bilateral stenosis or occlusion at the terminal portion of the internal carotid artery (ICA) and by an abnormal vascular network at the base of the brain<sup>1,3,10,14,16</sup>. There are two types of moyamoya disease: juvenile-onset and adult-onset<sup>1,3,7</sup>, and it has been suggested that these two types are actually clinically distinct entities<sup>7</sup>. Indirect bypass surgeries are known to be more effective for preventing recurrent attacks in children with juvenile-onset moyamoya disease than in adults with the hemorrhagic type<sup>1,3</sup>.

We present the case of a female patient with juvenile-onset moyamoya disease who experienced an intracranial hemorrhage long after successful revascularization surgery.

## CASE REPORT

A 17-year-old girl experienced a sudden mental change while she was running in a play ground. She presented with a drowsy mentality, irritability and left side motor weakness (grade IV). A brain computed tomographic scan on admission demonstrated an intracerebral hemorrhage in the right parietal lobe with an intraventricular hemorrhage (Fig. 1). External ventricular drainage was undertaken to resolve obstructive hydrocephalus and drain the hematoma. Ten years ago, the patient had a symptomatic left hemisphere from repeated transient ischemic attacks (TIAs), whereas the right hemisphere had never been symptomatic before. At that time, she underwent sequential right then left side encephaloduroarterio synangiosis (EDAS) and subsequently, she experienced no symptoms of TIA or hemorrhagic attack until this admission.

Magnetic resonance imaging (MRI), perfusion MRI and magnetic resonance (MR) angiography were performed, and revealed parietal lobe edema around the hematoma, but no perfusion defect and a good collateral circulation. In perfusion MRI, relatively high perfusion was noted in the territory of the right side middle cerebral artery (MCA), which implied the presence of hemodynamic stress (Fig. 2).

• Received : August 13, 2008 • Revised : March 26, 2009

• Accepted : August 17, 2009

• Address for reprints : Young Seok Park, M.D.

Department of Neurosurgery, Severance Children's Hospital, Brain Korea 21 Project for Medical Science, Brain Research Institute, Yonsei University College of Medicine, 250 Seongsan-ro, Seodaemun-gu, Seoul 120-752, Korea

Tel : +82-2-2228-2174, Fax : +82-2-393-9979

E-mail : ysparkmd@yuhs.ac

On the other hand, MR angiography revealed good revascularization of bilateral MCA territories, which could have been due to grafted superficial temporal arteries and middle meningeal arteries. Cerebral vessel angiography was also performed, and showed remaining small amount of right basal moyamoya vessels but no definite evidence of a microaneurysm. Collateral vessels from the right superficial temporal artery to the right MCA territory were well formed

(Fig. 3). The patient was discharged without any neurological deficit and complications.

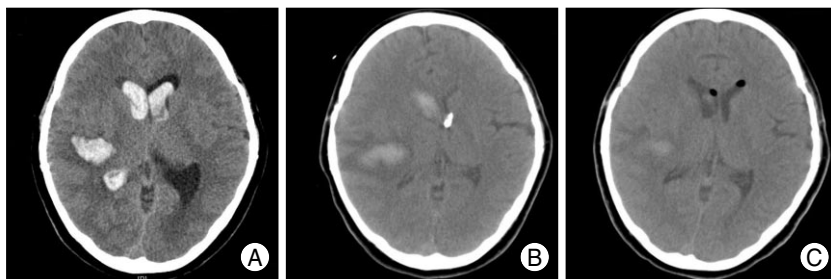
## DISCUSSION

In this case, we have shown that cerebral hemorrhage may occur long after good collateralization of indirect bypass surgery in moyamoya disease. Specifically, we have demon-

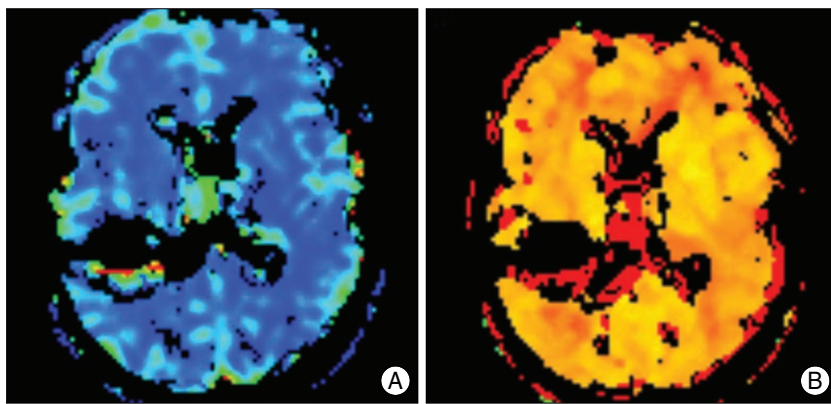
strated good collateralization by perfusion MRI and cerebral angiography. This case thus lends probabilities to amend our old interpretation that hemorrhage usually develop in poor collateralization.

Delayed hemorrhage long after successful revascularization surgery for juvenile-onset moyamoya disease is rare, but it has been reported. Three cases of juvenile-onset moyamoya disease were identified as delayed hemorrhage long after bypass surgery. There was one case of delayed intraventricular hemorrhage 5 years after indirect bypass surgery for juvenile-onset moyamoya disease<sup>12)</sup>. Another case, a 23-year-old female, showed delayed intracranial hemorrhage 16 years after undergoing bilateral EDAS due to TIA symptoms<sup>5)</sup>. There was another case of a 25-year-old female with hemorrhage who experienced TIAs 17 years ago and underwent bilateral direct bypass surgeries 16 years ago<sup>5)</sup>.

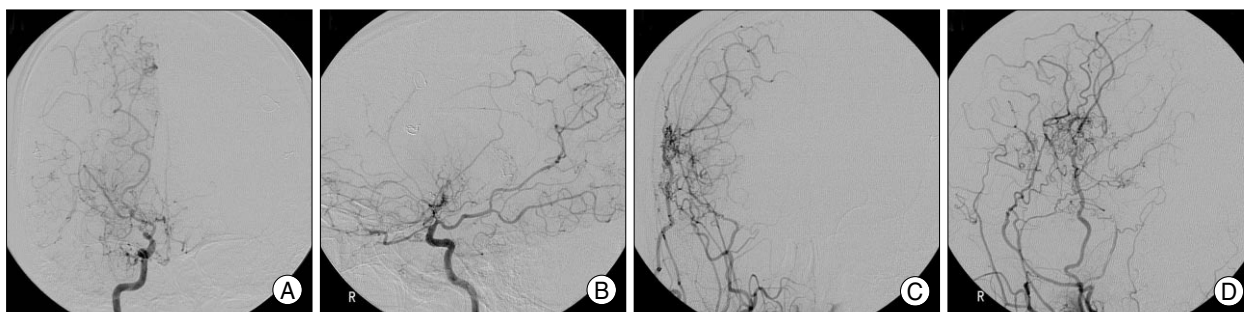
The typical symptoms of juvenile-onset moyamoya disease mainly involve repeated and progressive ischemic symptoms, while those of adult-



**Fig. 1.** A : Initial brain computed tomographic scan at this admission. Intraventricular and intracerebral hemorrhages on the right parietal lobe are visualized. Obstructive hydrocephalus is presented with obstruction of the third ventricular space by hematoma. B : Ten days later after external ventricular drainage, the right parietal lobe is edematous and midline shifting is presented. In addition, the intracerebral hematoma has resolved and the ventricle size has reduced as compared with the initial brain computed tomographic scan. C : Seventeen days later after operation, the ventricular catheter is removed. The image showing a midline shift improvement and a resolution of intraventricular and intracerebral hemorrhage. The ventricle size is returned normal.



**Fig. 2.** A : In perfusion magnetic resonance image of peak blood flow, no perfusion defect is observed in bilateral middle cerebral artery territories except the hematoma defect site. Right parietal lobe shows slightly higher peak blood flow near the hematoma. B : Image of blood volume, no blood volume reduction is observed in bilateral middle cerebral artery territories.



**Fig. 3.** Cerebral angiography reveals good collateral circulations from the superficial temporal artery to the cerebral cortex and a small amount of remained basal moyamoya vessels. A : Right internal carotid artery angiography in anteroposterior view. B : Right internal carotid artery angiography in lateral view. C : Right external carotid artery angiography in anteroposterior view. D : Right external carotid artery angiography in lateral view.

onset disease are mainly hemorrhagic and relatively stable<sup>1,2</sup>. About half of adult patients with moyamoya disease develop intracranial bleeding<sup>8</sup>. Even the good collateralization after indirect bypass, basal moyamoya vessels or hemodynamic stress can be aggravated. There are two main cause of intracranial bleeding in moyamoya disease; rupture of dilated, fragile moyamoya vessels or rupture of saccular aneurysm<sup>8</sup>.

Many reports have advocated that indirect revascularization surgeries are effective in preventing recurrent ischemic events in patients with juvenile-onset disease<sup>1-3</sup>. Ischemic symptoms do not always appear simultaneously in both hemispheres, there are some disagreements concerning the advisability of prophylactic bypass surgery on the asymptomatic side<sup>12</sup>. Bypass surgery on the asymptomatic hemisphere can be delayed until the development of ischemic symptoms, such as frequent TIAs<sup>12</sup>.

The right hemisphere of this patient did not show the common clinical course of juvenile-onset moyamoya disease. It seems to be progressed into hemorrhagic type. The right hemisphere did not suffer any clinical symptoms, such as TIAs or hemorrhage, until this event. Prophylactic revascularization surgery for this patient did not prevent hemorrhage in the right hemisphere, and hemorrhage occurred 10 years after bypass surgery. The clinical course of the disease resembled that of adult-onset moyamoya disease.

The proposed mechanisms of hemorrhage in moyamoya disease are as follows. Impairment of the hemodynamic reserves required to change the intracranial perfusion pressure and failure of moyamoya blood vessels due to elevated hemodynamic stress could result in hemorrhage<sup>11,15</sup>. Microaneurysms in moyamoya vessels could also act as sources of cerebral hemorrhage<sup>14</sup>. However, in the majority of cases, recurrent hemorrhages do not result from breakdown at specific weak points, but are rather caused by diffuse vulnerabilities of collateral vessels adjacent to the lateral ventricle<sup>7</sup>. Gradient-echo T2-weighted 3-T MRI is a good tool for detecting microbleeds, which are regarded as a general marker of microvascular vulnerability and are thus a risk factor for hemorrhage<sup>5</sup>.

Although the precise mechanism of hemorrhage in moyamoya disease remains unknown and no definite preventive treatment has been devised<sup>6</sup>, researchers continue to find better ways of preventing hemorrhage<sup>5-7,10,13,15</sup>. Many reports have addressed treatment options for reducing the rates of re-bleeding in hemorrhagic moyamoya disease<sup>4,10,13,15,16</sup>; however, reductions in re-bleeding incidents in hemorrhagic moyamoya patients after surgical treatment are difficult to assess due to a lack of long-term postoperative follow-up data<sup>14,15</sup>. Nevertheless, bypass surgery is a

powerful tool in this area because it likely aids in the prevention of re-bleeding, which increases the blood flow in revascularized collateral vessels and reduces the number of moyamoya vessels<sup>10,13</sup>.

The present case is very rare, but it leads us to speculate that abnormal moyamoya vessels may progress even after successful surgery. It could be possible that remained abnormal vasculature in the asymptomatic side could be a reason of hemorrhage.

Long-term follow-up studies would be needed in order to establish a natural history of moyamoya disease. A prospective randomized controlled trial is now underway in Japan in order to determine the effect of bilateral direct revascularization surgery on patients with adult hemorrhagic moyamoya disease<sup>9,14</sup>. Through these efforts, further development on the integration of knowledge of moyamoya disease would make it possible for clinicians to determine the best strategies for the treatment and prevention of moyamoya disease.

## CONCLUSION

Hemorrhagic events can occur long after successful EDAS in juvenile-onset moyamoya disease, and thus a lifetime follow-up strategy might be necessary even if a good collateral circulation has been established. Good collateralization of cerebral angiography or perfusion MRI after indirect bypass surgery would ensure against ischemic symptoms, not a hemorrhage.

## References

1. Choi JU, Kim DS, Kim EY, Lee KC : Natural history of moyamoya disease: comparison of activity of daily living in surgery and non surgery groups. *Clin Neurol Neurosurg* 99 Suppl 2 : S11-S18, 1997
2. Fung LW, Thompson D, Ganesan V : Revascularisation surgery for paediatric moyamoya : a review of the literature. *Childs Nerv Syst* 21 : 358-364, 2005
3. Isono M, Ishii K, Kamida T, Inoue R, Fujiki M, Kobayashi H : Long-term outcomes of pediatric moyamoya disease treated by encephalo-duro-arterio-synangiosis. *Pediatr Neurosurg* 36 : 14-21, 2002
4. Kawaguchi S, Sakaki T, Kakizaki T, Kamada K, Shimomura T, Iwanaga H : Clinical features of the haemorrhage type moyamoya disease based on 31 cases. *Acta Neurochir (Wien)* 138 : 1200-1210, 1996
5. Kikuta K, Takagi Y, Nozaki K, Hanakawa T, Okada T, Mikuni N, et al. : Asymptomatic microbleeds in moyamoya disease : T2\*-weighted gradient-echo magnetic resonance imaging study. *J Neurosurg* 102 : 470-475, 2005
6. Kikuta K, Takagi Y, Nozaki K, Sawamoto N, Fukuyama H, Hashimoto N : The presence of multiple microbleeds as a predictor of subsequent cerebral hemorrhage in patients with moyamoya disease. *Neurosurgery* 62 : 104-111; discussion 111-112, 2008
7. Kobayashi E, Saeki N, Oishi H, Hirai S, Yamaura A : Long-term natural history of hemorrhagic moyamoya disease in 42 patients. *J*

- Neurosurg 93 : 976-980, 2000
8. Kuroda S, Houkin K : Moyamoya disease : current concepts and future perspectives. *Lancet Neurol* 7 : 1056-1066, 2008
  9. Miyamoto S; Japan Adult Moyamoya Trial Group : Study design for a prospective randomized trial of extracranial-intracranial bypass surgery for adults with moyamoya disease and hemorrhagic onset-the Japan Adult Moyamoya Trial Group. *Neurol Med Chir (Tokyo)* 44 : 218-219, 2004
  10. Morioka M, Hamada J, Todaka T, Yano S, Kai Y, Ushio Y : High-risk age for rebleeding in patients with hemorrhagic moyamoya disease : long-term follow-up study. *Neurosurgery* 52 : 1049-1054; discussion 1054-1055, 2003
  11. Nagasaka T, Hayashi S, Naito T, Okamoto T, Ikeda H, Inao S : Concomitant cerebral infarction and intraventricular hemorrhage in moyamoya disease. Case report. *J Neurosurg* 106 (5 Suppl) : 388-390, 2007
  12. Nagata S, Matsushima T, Morioka T, Matsukado K, Mihara F, Sasaki T, et al. : Unilaterally symptomatic moyamoya disease in children : long-term follow-up of 20 patients. *Neurosurgery* 59 : 830-836; discussion 836-837, 2006
  13. Okada Y, Shima T, Nishida M, Yamane K, Yamada T, Yamanaka C : Effectiveness of superficial temporal artery-middle cerebral artery anastomosis in adult moyamoya disease : cerebral hemodynamics and clinical course in ischemic and hemorrhagic varieties. *Stroke* 29 : 625-630, 1998
  14. Otawara Y, Ogasawara K, Seki K, Kibe M, Kubo Y, Ogawa A : Intracerebral hemorrhage after prophylactic revascularization in a patient with adult moyamoya disease. *Surg Neurol* 68 : 335-337; discussion 337, 2007
  15. Yoshida Y, Yoshimoto T, Shirane R, Sakurai Y : Clinical course, surgical management, and long-term outcome of moyamoya patients with rebleeding after an episode of intracerebral hemorrhage : an extensive follow-up study. *Stroke* 30 : 2272-2276, 1999
  16. Zipfel GJ, Fox DJ Jr, Rivet DJ : Moyamoya disease in adults: the role of cerebral revascularization. *Skull Base* 15 : 27-41, 2005